



General

Guideline Title

An official ATS/ERS/JRS/ALAT clinical practice guideline: treatment of idiopathic pulmonary fibrosis. An update of the 2011 clinical practice guideline.

Bibliographic Source(s)

Raghu G, Rochwerf B, Zhang Y, Cuello Garcia CA, Azuma A, Behr J, Brozek JL, Collard HR, Cunningham W, Homma S, Johkoh T, Martinez FJ, Myers J, Protzko SL, Richeldi L, Rind D, Selman M, Theodore A, Wells AU, Hoogsteden H, Schunemann HJ, ATS, ERS, JRS, and ALAT. An official ATS/ERS/JRS/ALAT clinical practice guideline: treatment of idiopathic pulmonary fibrosis. An update of the 2011 clinical practice guideline. *Am J Respir Crit Care Med*. 2015 Jul 15;192(2):e3-19. [70 references] [PubMed](#)

Guideline Status

This is the current release of the guideline.

This guideline meets NGC's 2013 (revised) inclusion criteria.

Recommendations

Major Recommendations

The strength of the recommendations (strong, conditional) and levels of evidence (high, moderate, low or very low) are defined at the end of the "Major Recommendations" field.

Note: See Table 2 in the original guideline document for comparison of recommendations in the 2015 and 2011 Idiopathic Pulmonary Fibrosis Guidelines.

Recommendations for Specific Treatment Questions

Please see the online supplement (see the "Availability of Companion Documents" field), which includes supporting evidence profiles for each recommendation.

Question 1: Should patients with idiopathic pulmonary fibrosis (IPF) be treated with anticoagulation?

Recommendation

The committee recommends that clinicians not use warfarin anticoagulation in patients with IPF who do not have a known alternative indication for its use (strong recommendation against, low confidence in estimates of effect).

Question 2: Should patients with IPF be treated with imatinib, a tyrosine kinase inhibitor?

Recommendation

The committee recommends that clinicians not use imatinib in patients with IPF (strong recommendation, moderate confidence in estimates of effect).

Question 3: Should patients with IPF be treated with combination prednisone, azathioprine, and *N*-acetylcysteine?

Recommendation

The committee recommends that clinicians not use the combination therapy of *N*-acetylcysteine, azathioprine, and prednisone in patients with IPF (strong recommendation, low confidence in estimates of effect).

Question 4: Should patients with IPF be treated with ambrisentan, a selective ER-A endothelin receptor antagonist?

Recommendation

The committee recommends that clinicians not use ambrisentan, a selective ER-A endothelin receptor antagonist, in patients with IPF, regardless of the presence or absence of pulmonary hypertension (PH) (strong recommendation against, low confidence in estimates of effect).

Question 5: Should Patients with IPF be treated with nintedanib, a tyrosine kinase inhibitor?

Recommendation

The committee suggests that clinicians use nintedanib in patients with IPF (conditional recommendation, moderate confidence in estimates of effect).

Question 6: Should patients with IPF be treated with pirfenidone?

Recommendation

The committee suggests that clinicians use pirfenidone in patients with IPF (conditional recommendation, moderate confidence in estimates of effect).

Question 7: Should patients with IPF be treated with antacid medication?

Recommendation

The committee suggests that clinicians use regular antacid treatment for patients with IPF (conditional recommendation, very low confidence in estimates of effect).

Question 8: Should patients with IPF be treated with sildenafil, a phosphodiesterase-5 inhibitor?

Recommendation

The committee suggests that clinicians not use sildenafil, a phosphodiesterase-5 inhibitor, for treatment of IPF (conditional recommendation against, moderate confidence in estimates of effect).

Question 9: Should patients with IPF be treated with bosentan or macitentan, dual endothelin receptor antagonists (ER-A and ER-B)?

Recommendation

The committee suggests that clinicians not use bosentan or macitentan, both dual ER-A and ER-B endothelin receptor antagonists, for the treatment of IPF (conditional recommendation against, low confidence in estimates of effect).

Question 10: Should patients with IPF be treated with *N*-acetylcysteine monotherapy?

Recommendation

The committee suggests that clinicians not use *N*-acetylcysteine monotherapy in patients with IPF (conditional recommendation, low confidence in estimates of effect).

Question 11: Should patients with IPF be treated with bilateral lung transplantation versus single-lung transplantation?

Recommendation

The committee did not make a recommendation regarding single versus bilateral lung transplantation in patients with IPF.

Question 12: Should PH be treated in patients with IPF?

Recommendation

The committee did not make a recommendation regarding treatment of PH in patients with IPF.

Definitions

Quality of the Evidence and Implications

Quality of Evidence (GRADE)	The quality of evidence is a judgement about the extent to which the authors can be confident that the estimates of effects are correct. This judgement is made using the Grading of Recommendations Assessment, Development and Evaluation (GRADE) system, and are provided for each outcome. The judgements are based on the type of study design (randomized trials versus observational studies), the risk of bias, the consistency of the results across studies, and the precision of the overall estimates across studies. For each outcome, the quality of the evidence is rated as high, moderate, low, and very low using the following definitions:
High ++++	Further research is very unlikely to change confidence in the estimate of effect.
Moderate +++O	Further research is likely to have an important impact on confidence in the estimate of effect and may change the estimate.
Low ++OO	Further research is very likely to have an important impact on confidence in the estimate of effect and is likely to change the estimate.
Very low +OOO	The authors are very uncertain about the estimate. (For more information about the GRADE system see www.gradeworkinggroup.org <input type="text"/>).

Interpretation of Strong and Conditional Recommendations for Stakeholders (Patients, Clinicians, and Health Care Policy Makers)

Implications for:	Strong Recommendation	Conditional Recommendation
Patients	Most individuals in this situation would want the recommended course of action, and only a small proportion would not.	The majority of individuals in this situation would want the suggested course of action, but many would not.
Clinicians	Most individuals should receive the intervention. Adherence to this recommendation according to the guideline could be used as a quality criterion or performance indicator. Formal decision aids are not likely to be needed to help individuals make decisions consistent with their values and preferences.	Recognize that different choices will be appropriate for individual patients and that you must help each patient arrive at a management decision consistent with his or her values and preferences. Decision aids may be useful in helping individuals to make decisions consistent with their values and preferences.
Policy Makers	The recommendation can be adopted as policy in most situations.	Policy making will require substantial debate and involvement of various stakeholders.

Clinical Algorithm(s)

None provided

Scope

Disease/Condition(s)

Idiopathic pulmonary fibrosis (IPF)

Guideline Category

Management

Treatment

Clinical Specialty

Critical Care

Internal Medicine

Pulmonary Medicine

Thoracic Surgery

Intended Users

Advanced Practice Nurses

Nurses

Physician Assistants

Physicians

Guideline Objective(s)

To analyze evidence reported since publication of the prior guideline in 2011 and to update the treatment recommendations accordingly

Target Population

Patients with idiopathic pulmonary fibrosis (IPF)

Interventions and Practices Considered

1. Nintedanib
2. Pirfenidone
3. Antacid therapy

Note: The following interventions were considered but not recommended:

- Anticoagulation (warfarin)
- Imatinib
- Combination prednisone, azathioprine and *N*-acetylcysteine
- Selective endothelin receptor antagonist (ambrisentan)
- Dual endothelin receptor antagonists (macitentan, bosentan)
- Phosphodiesterase-5 inhibitor (sildenafil)
- *N*-acetylcysteine monotherapy
- Single versus bilateral lung transplantation

Major Outcomes Considered

- Mortality
- Disease progression (defined as worsening pulmonary function test [PFT] results, progressive fibrosis on high-resolution computed tomography scan, acute respiratory decline, death, changes over time in forced vital capacity [FVC] or diffusing capacity of the lung for carbon monoxide [DL_{CO}])

- Acute exacerbation
- Adverse events
- Serious adverse events
- Quality of life
- Short of breath/Borg dyspnea score
- Oxygen saturation
- Function (measured with change in 6-minute walk distance)

Methodology

Methods Used to Collect/Select the Evidence

Searches of Electronic Databases

Description of Methods Used to Collect/Select the Evidence

Literature Search

In collaboration with the Methods Group (MG), an information scientist designed a search strategy using medical subject heading keywords and text words (see the online supplement [see the "Availability of Companion Documents" field]) limited to human studies or nonindexed citations and articles in English or in any language with English abstracts. The Ovid platform was used to search MEDLINE, EMBASE, Cochrane Registry of Controlled Trials, Health Technology Assessment, and the Database of Abstracts of Reviews of Effects for May 2010 through May 2014. An update was performed in June 2014, immediately before the meeting at McMaster University. Reviewers contacted experts and reviewed previous meta-analyses for additional articles. The search retrieved 9,663 citations, minus duplicates. On the basis of predefined eligibility criteria, 54 citations were included for full text review, of which 34 were excluded with reasons and 20 were included in the evidence update (see the online supplement).

Committee members were also queried for any additional studies not identified by the search. If adequate outcome data were not available from randomized controlled trials (RCTs), observational studies were also used to support recommendations.

Two reviewers from the MG screened titles and abstracts to identify articles for full review and evaluated the full text of articles deemed potentially relevant by either reviewer. Disagreement was resolved by consensus among the MG group.

Number of Source Documents

Twenty citations were included in the evidence update (see the flow chart of search results in the online supplement [see the "Availability of Companion Documents" field]).

Methods Used to Assess the Quality and Strength of the Evidence

Weighting According to a Rating Scheme (Scheme Given)

Rating Scheme for the Strength of the Evidence

Quality of the Evidence and Implications

Quality of Evidence (GRADE)	The quality of evidence is a judgement about the extent to which the authors can be confident that the estimates of effects are correct. This judgements are made using the Grading of Recommendations Assessment, Development and Evaluation (GRADE) system, and are provided for each outcome. The judgements are based on the type of study design (randomized trials versus observational studies), the risk of bias, the consistency of the results across studies, and the precision of the overall estimates across studies. For each outcome, the quality of the evidence is rated as high, moderate, low, and very low using the following definitions:
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High ++++	Further research is very unlikely to change confidence in the estimate of effect.
Moderate +++O	Further research is likely to have an important impact on confidence in the estimate of effect and may change the estimate.
Low ++OO	Further research is very likely to have an important impact on confidence in the estimate of effect and is likely to change the estimate.
Very low +OOO	The authors are very uncertain about the estimate. (For more information about the GRADE system see www.gradeworkinggroup.org <input type="text"/>).

Methods Used to Analyze the Evidence

Meta-Analysis

Review of Published Meta-Analyses

Systematic Review with Evidence Tables

Description of the Methods Used to Analyze the Evidence

Evidence Review

Evidence summaries for each question were prepared by the McMaster methodology team, following the Grading of Recommendations Assessment, Development and Evaluation (GRADE) approach, using the GRADEpro Guideline Development Tool online software. All committee members reviewed the summaries of evidence, and corrections were made when appropriate. The committee based the evidence on the 2011 evidence summaries that had been produced for that document. These summaries were updated, if necessary, with additional recent randomized controlled trials (RCTs).

Data abstraction occurred in duplicate, using predesigned data abstraction forms that had been piloted before being used. In addition to clinical data, individual study risk of bias was assessed independently by two reviewers, using the Cochrane Risk of Bias tool for RCTs and the Ottawa-Newcastle tool for observational studies.

Results from identified studies with the same treatment agent were pooled, and meta-analyses, using the Cochrane Collaboration Review Manager, version 5.2, were reviewed. Pooling and meta-analyses of study data were independently performed by the Methods Group (MG) specifically for this guideline document. All data fulfilling the *a priori* inclusion criteria were included, and pooled analysis presented in this document may at times differ from other published meta-analyses, depending on inclusion or exclusion criteria. Subsequently, the overall certainty in effect estimates (also known as confidence in effect estimate) for each outcome of interest was assessed following the GRADE approach, based on the following criteria: risk of bias, precision, consistency, directness of the evidence, risk for publication bias, presence of dose-effect relationship, magnitude of effect, and assessment of the effect of plausible residual confounding or bias. The confidence in effect estimates for each outcome was categorized into one of four levels: high, moderate, low, or very low (see the "Rating Scheme for the Strength of the Evidence" field).

Methods Used to Formulate the Recommendations

Expert Consensus

Description of Methods Used to Formulate the Recommendations

Committee Composition

This guideline was developed by a multidisciplinary committee that consisted of 8 pulmonologists with recognized idiopathic pulmonary fibrosis (IPF) expertise, 3 general pulmonologists, a pulmonologist-methodologist, an allergist-methodologist, a general internist, a chest radiologist, a pulmonary pathologist, an information scientist, and a patient with IPF, who was recommended for participation by the Coalition for Pulmonary Fibrosis and was not known to any of the committee members. The committee included a chair, two co-chairs, and committee members representing the American Thoracic Society (ATS), European Respiratory Society (ERS), Japanese Respiratory Society (JRS) and the Latin

American Thoracic Association (ALAT).

The committee worked with the Methods Group (MG), which comprised five health research methodologists from the MacGRADE Centre at McMaster University who had expertise in evidence synthesis and the guideline development process. Four of these methodologists are also clinicians. The MG conducted systematic reviews and prepared the systematic evidence summaries following the Grading of Recommendations Assessment, Development and Evaluation (GRADE) approach.

Meetings

Face-to-face planning meetings were held during the 2013 ATS International Conference in Philadelphia, Pennsylvania, at which the committee discussed the scope and objectives of the project, and during the 2014 ATS International Conference in San Diego, California, to go over the proceedings of the upcoming face-to-face meeting in June 2014 in Hamilton, Ontario, Canada. Members who could not attend the actual face-to-face meetings participated in person live by teleconference. Additional planning meetings were held regularly over telephone between the committee chair, the co-chair, and the MG. Conference calls and email correspondence were used to discuss specific issues requiring input from others.

The entire guideline committee met at the McMaster Health Forum in Hamilton, Ontario, Canada, on June 9–10, 2014, at which the evidence summaries were presented and discussed, and the recommendations were formulated. Three members participated through teleconference and webinar. The methodologists took notes of all matters and points discussed and documented all the recommendations and proceedings.

Two follow-up teleconference webinars were held on June 23 and July 15, 2014, to complete the guideline development for two of the 12 treatment questions (questions on single versus bilateral lung transplantation and treatment of IPF-associated pulmonary hypertension [PH]). Three members were not able to participate live during the first teleconference-webinar, and five members were not able to join the second teleconference-webinar, but all provided feedback and discussion via emails. All meetings were attended by staff from the ATS documents unit.

Formulating Clinical Questions

The committee used the treatment section of the 2011 guideline document as a starting point. Twelve specific questions pertinent and relevant to current clinical practice were addressed to update the recommendations pertinent to treatment of IPF. Most of these questions were previously addressed, and formal recommendations had been provided in the 2011 document. Questions pertinent to the management of patients with IPF with pulmonary rehabilitation, oxygen supplementation, antibiotics, palliative care, mechanical ventilation, and specific questions that had received a "strong against" or "strong for" in the 2011 guideline were not readdressed in this update unless the literature search revealed new and pertinent evidence.

The committee selected outcomes of interest for each question, using the 2011 document as a guide in addition to following the approach suggested by the GRADE working group. All outcomes were identified *a priori*, and the committee explicitly rated their relative importance (from the perspective of a patient with IPF) from not important to critical. Rankings of all outcomes were agreed on through consensus of the committee.

Development of Clinical Recommendations

The committee developed recommendations based on the GRADE evidence profiles for each recommendation. The committee employed the GRADE evidence to decision frameworks in the guideline development tool to help organize discussion around each recommendation and ensure each of the following factors was considered in recommendation development: the quality of the evidence, the balance of desirable and undesirable consequences of compared management options, the assumptions about the values and preferences associated with the decision, the implications for resource use and health equity, the acceptability of intervention to stakeholders, and the feasibility of implementation (see the evidence to decision frameworks document [see the "Availability of Companion Documents" field]). Recommendations and their strength were decided by consensus, and only one recommendation required voting because of inability to achieve consensus. The committee agreed on the final wording of recommendations and remarks with further qualifications for each recommendation (e.g., subgroup considerations, justification, implementation considerations).

The recommendations were either "strong" or "conditional," according to the GRADE approach. Conditional recommendations are synonymous with weak recommendations (see the "Rating Scheme for the Strength of the Recommendations" field). The 2011 guideline had used the nomenclature "weak," but to improve clarity (which conditions are relevant to implement the recommendation) and facilitate translation of guidelines to other languages, GRADE uses the term "conditional" as an alternative. Factors influencing the strength of the recommendation include the strength of evidence, the outcomes studies and associated importance to patients, the desirable and undesirable consequences of treatment, the cost of treatment, the implications of treatment on health equity, the feasibility of treatment, the acceptability of treatment to important stakeholders, and potential treatment monitoring and implementation issues.

As suggested by GRADE, the committee used the phrasing "we recommend" for strong recommendations and "we suggest" for conditional

recommendations. The "Rating Scheme for the Strength of the Recommendations" field provides suggested interpretation of these recommendations by intended stakeholders, including patients, clinicians, and health policy makers. For two questions, the panel decided to not offer a recommendation because it was realized that additional evidence, mostly indirect and resource- or cost-related, should be considered to fully inform the panel, and the panel documented this as "no recommendation."

There are two important aspects of the recommendations to consider. First, recommendations of similar strength should not be interpreted as equivalent recommendations. Each recommendation's strength is the net result of considering the multiple factors described earlier, and therefore there may be different reasons that two recommendations are rated with the same strength (e.g., one recommendation may be conditional because it is based on very low confidence in effect estimates, whereas another recommendation may be conditional because the cost is so high that it is unclear that the potential benefits outweigh those costs for every patient). Second, the methodology used in making recommendations for or against the use of therapies in guidelines considers additional factors than those used by regulatory agencies (whose purpose is to review data submitted to them and subsequently consider approval of new treatments for use in patients).

Manuscript Preparation

The writing committee drafted the guideline document. The manuscript was then reviewed by the entire committee. Feedback was provided primarily by electronic communication and, to a lesser extent, during a face-to-face meeting at the European Respiratory Society (ERS) Congress on September 7, 2014, that included some of the committee members.

The entire committee (both conflicted and nonconflicted members) had the opportunity to correct factual errors, clarify the presentation of background information or evidence summaries, and suggest changes to the rationale sections if they improperly captured the discussion from the face-to-face meetings. However, only the nonconflicted voting members were permitted to comment on the recommendations. The conflicted chair and conflicted committee members were not permitted to comment on the recommendations and restricted their feedback to the presentation of the evidence and the identification of errors. The wording of recommendations (including strength and direction) was not altered once they were finalized by the nonconflicted members during the face-to-face meeting and teleconferences. One of the nonconflicted co-chairs confirmed that the written version of the guideline reflected the recommendations made by the nonconflicted members. The same process was followed for each version of the document.

Rating Scheme for the Strength of the Recommendations

Interpretation of Strong and Conditional Recommendations for Stakeholders (Patients, Clinicians, and Health Care Policy Makers)

Implications for:	Strong Recommendation	Conditional Recommendation
Patients	Most individuals in this situation would want the recommended course of action, and only a small proportion would not.	The majority of individuals in this situation would want the suggested course of action, but many would not.
Clinicians	Most individuals should receive the intervention. Adherence to this recommendation according to the guideline could be used as a quality criterion or performance indicator. Formal decision aids are not likely to be needed to help individuals make decisions consistent with their values and preferences.	Recognize that different choices will be appropriate for individual patients and that you must help each patient arrive at a management decision consistent with his or her values and preferences. Decision aids may be useful in helping individuals to make decisions consistent with their values and preferences.
Policy Makers	The recommendation can be adopted as policy in most situations.	Policy making will require substantial debate and involvement of various stakeholders.

Cost Analysis

A formal cost analysis was not performed and published cost analyses were not reviewed.

Method of Guideline Validation

Internal Peer Review

Description of Method of Guideline Validation

The writing committee drafted the guideline document. The manuscript was then reviewed by the entire committee. Feedback was provided primarily by electronic communication and, to a lesser extent, during a face-to-face meeting at the European Respiratory Society (ERS) Congress on September 7, 2014, that included some of the committee members.

The final approved version was submitted to each cosponsoring professional society for peer review.

This official clinical practice guideline of the American Thoracic Society (ATS) was approved by the ATS, May 2015, the ERS, April 2015, the Japanese Respiratory Society (JRS), April 2015, and the Latin American Thoracic Association (ALAT), April 2015.

Evidence Supporting the Recommendations

Type of Evidence Supporting the Recommendations

The type of supporting evidence is identified and graded for each recommendation (see the "Major Recommendations" field).

Benefits/Harms of Implementing the Guideline Recommendations

Potential Benefits

The guideline should empower clinicians to interpret these recommendations in the context of individual patient values and preferences and to make appropriate clinical decisions about treatment of patients with idiopathic pulmonary fibrosis (IPF). See the "Summary of the Evidence" and the "Justification and Implementation Considerations" sections in the original guideline document for discussions of potential benefits of individual recommendations.

Potential Harms

See the "Summary of the Evidence" and the "Justification and Implementation Considerations" sections in the original guideline document for discussions of potential harms of individual recommendations.

Qualifying Statements

Qualifying Statements

- For each recommendation, it is important to consider both the summary of evidence reviewed and discussed by the nonconflicted members of the committee and remarks for each specific treatment question, including the values and preferences, before applying these recommendations to specific clinical situations or policy decisions.
- Clinicians, patients, third-party payers, and other stakeholders should never view these recommendations as dictates. No guideline or recommendations can take into account all of the often compelling unique individual clinical circumstances. Therefore, no one charged with evaluating clinicians' actions should attempt to apply the recommendations from this guideline by rote or in a blanket fashion.
- This guideline does not provide recommendations for one treatment regimen over another. With the exception of the recommendation against using prednisone with azathioprine and *N*-acetylcysteine, the guideline does not provide suggestions for or against combination regimens or sequential therapies. Therefore, the strong or conditional rating for each recommendation must be weighed individually (i.e., two recommendations with the same strong or conditional rating should not by default be considered equivalent recommendations), factoring in all components used to determine the grade of the recommendation, including the confidence in effect estimates, outcomes studies, desirable and undesirable consequences of treatment, cost of treatment, implications of treatment on health equity, and feasibility of treatment. The methods used by guideline panels to appraise the evidence are different than those used by regulatory agencies when they review applications seeking market approval for the use of pharmacologic agents for treatment of idiopathic pulmonary fibrosis (IPF).

Implementation of the Guideline

Description of Implementation Strategy

An implementation strategy was not provided.

Implementation Tools

Patient Resources

Quick Reference Guides/Physician Guides

For information about availability, see the *Availability of Companion Documents* and *Patient Resources* fields below.

Institute of Medicine (IOM) National Healthcare Quality Report Categories

IOM Care Need

Getting Better

Living with Illness

IOM Domain

Effectiveness

Identifying Information and Availability

Bibliographic Source(s)

Raghu G, Rochwerf B, Zhang Y, Cuello Garcia CA, Azuma A, Behr J, Brozek JL, Collard HR, Cunningham W, Homma S, Johkoh T, Martinez FJ, Myers J, Protzko SL, Richeldi L, Rind D, Selman M, Theodore A, Wells AU, Hoogsteden H, Schunemann HJ, ATS, ERS, JRS, and ALAT. An official ATS/ERS/JRS/ALAT clinical practice guideline: treatment of idiopathic pulmonary fibrosis. An update of the 2011 clinical practice guideline. *Am J Respir Crit Care Med*. 2015 Jul 15;192(2):e3-19. [70 references] [PubMed](#)

Adaptation

Not applicable: The guideline was not adapted from another source.

Date Released

2015 Jul 15

Guideline Developer(s)

American Thoracic Society - Medical Specialty Society

European Respiratory Society - Professional Association

Japanese Respiratory Society - Medical Specialty Society

Latin American Thoracic Association - Medical Specialty Society

Source(s) of Funding

McMaster University provided meeting facilities and logistical support, and the sponsoring societies provided the financial support for expenses resulting from the meeting and conference calls. The views and interests of the American Thoracic Society (ATS), the European Respiratory Society (ERS), the Japanese Respiratory Society (JRS), and the Latin American Thoracic Association (ALAT), as well as of any commercial entity that provided external funding for professional societies, had no influence on the topics discussed and recommendations made.

Guideline Committee

ATS/ERS/JRS/ALAT Committee on Treatment of IPF

Composition of Group That Authored the Guideline

Committee Members: Ganesh Raghu, M.D. (*Chair*); Henk Hoogsteden, MD (*Co-chair*); Holger J. Schünemann, MD, PhD (*Co-chair*); Bram Rochwerf, MD, MSc; Yuan Zhang, MSc; Carlos A. Cuello Garcia, MD, MSc; Arata Azuma, MD, PhD; Juergen Behr, MD; Jan L. Brozek, MD, PhD; Harold R. Collard, MD; William Cunningham*; Sakae Homma, MD; Takeshi Johkoh, MD; Fernando J. Martinez, MD, MS; Jeffrey Myers, MD; Shandra L. Protzko; Luca Richeldi, MD, PhD; David Rind, MD; Moisés Selman, MD; Arthur Theodore, MD; Athol U. Wells, MD

*Deceased

Financial Disclosures/Conflicts of Interest

Confidentiality Agreement and Conflict-of-interest Management

Committee members signed a confidentiality agreement and disclosed all potential conflicts of interest according to the American Thoracic Society (ATS) and European Respiratory Society (ERS) policies. Two of the co-chairs (G.R. and H.J.S.) reviewed all potential conflicts of interest of committee members with the staff of the ATS conflict-of-interest and documents units.

All of the eight pulmonologists with recognized idiopathic pulmonary fibrosis (IPF) expertise were considered to either have major financial or intellectual conflicts based on disclosures or participation in IPF clinical trials/studies; although they were permitted to participate in the discussions of the evidence with the rest of the committee, they were instructed to abstain from discussions related to the evidence to decision framework, formulating and grading recommendations, and voting on recommendations if necessary. This approach was applied to all questions, not just those in which they had a perceived conflict of interest. Conflicted members were allowed to stay in the same room while discussions among nonconflicted members took place to provide expert input; however, they could do so only when specifically requested by nonconflicted members. Adherence to the rules was strict, with one of the co-chairs responsible for monitoring the discussions for adherence to these rules.

The remaining nine nonconflicted committee members were allowed unrestricted participation. Two of the voting members were members of the Methods Group (MG); they are clinicians with extensive expertise in the guideline development process. The rest of the MG and the librarian also participated in discussions, but were nonvoting participants.

Specific author disclosures are provided in the original guideline document.

Guideline Status

This is the current release of the guideline.

This guideline meets NGC's 2013 (revised) inclusion criteria.

Guideline Availability

Available from the [American Thoracic Society \(ATS\) Web site](#) .

Availability of Companion Documents

The following are available:

- Raghu G, Rochwerg B, Zhang Y, Cuello Garcia CA, Azuma A, Behr J, Brozek JL, Collard HR, Cunningham W, Homma S, Johkoh T, Martinez FJ, Myers J, Protzko SL, Richeldi L, Rind D, Selman M, Theodore A, Wells AU, Hoogsteden H, Schunemann HJ, ATS, ERS, JRS, and ALAT. An official ATS/ERS/JRS/ALAT clinical practice guideline: treatment of idiopathic pulmonary fibrosis. An update of the 2011 clinical practice guideline. Executive summary. *Am J Respir Crit Care Med*. 2015 Jul 15;192(2):238-248. Available from the [American Thoracic Society \(ATS\) Web site](#) .
- Raghu G, Rochwerg B, Zhang Y, Cuello Garcia CA, Azuma A, Behr J, Brozek JL, Collard HR, Cunningham W, Homma S, Johkoh T, Martinez FJ, Myers J, Protzko SL, Richeldi L, Rind D, Selman M, Theodore A, Wells AU, Hoogsteden H, Schunemann HJ, ATS, ERS, JRS, and ALAT. An official ATS/ERS/JRS/ALAT clinical practice guideline: treatment of idiopathic pulmonary fibrosis. An update of the 2011 clinical practice guideline. Online supplement. 2015 Jul. 24 p. Available from the [ATS Web site](#) .
- Raghu G, Rochwerg B, Zhang Y, Cuello Garcia CA, Azuma A, Behr J, Brozek JL, Collard HR, Cunningham W, Homma S, Johkoh T, Martinez FJ, Myers J, Protzko SL, Richeldi L, Rind D, Selman M, Theodore A, Wells AU, Hoogsteden H, Schunemann HJ, ATS, ERS, JRS, and ALAT. An official ATS/ERS/JRS/ALAT clinical practice guideline: treatment of idiopathic pulmonary fibrosis. An update of the 2011 clinical practice guideline. Evidence to decision frameworks. 2015 Jul. 55 p. Available from the [ATS Web site](#) .

Patient Resources

The following is available:

- Patient Information Series. Idiopathic pulmonary fibrosis (IPF). 2015 Mar. 2 p. Available from the [American Thoracic Society \(ATS\) Web site](#) .

Please note: This patient information is intended to provide health professionals with information to share with their patients to help them better understand their health and their diagnosed disorders. By providing access to this patient information, it is not the intention of NGC to provide specific medical advice for particular patients. Rather we urge patients and their representatives to review this material and then to consult with a licensed health professional for evaluation of treatment options suitable for them as well as for diagnosis and answers to their personal medical questions. This patient information has been derived and prepared from a guideline for health care professionals included on NGC by the authors or publishers of that original guideline. The patient information is not reviewed by NGC to establish whether or not it accurately reflects the original guideline's content.

NGC Status

This NGC summary was completed by ECRI Institute on November 11, 2015. The information was not verified by the guideline developer.

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